NEW YORK NEUROLOGICAL SOCIETY.

Stated Meeting, November 2d, 1897.

Dr. C. A. Herter, the Vice-president, in the chair.

THOMSEN'S DISEASE.

Dr. George W. Jacoby exhibited a typical case of Thomsen's disease. The patient, a young man, had been referred to him by Dr. Schwinn, of West Virginia, with a correct diagnosis. The patient was twenty-eight years of age and had lived in this country since 1884. There was nothing in the family history bearing upon the condition especially, except that a distant cousin was said to have walked stiffly and in a peculiar manner for fifteen years. The patient himself had always been delicate, but had been as active as other boys. He had had typhoid fever in 1889, and on recovering from this had first noticed a cramp in the legs. After a little it was found that he could not execute movements as quickly as before. In 1893, he first sought treatment. For the past year or two his arms and hands had also been affected. The condition varied considerably at different times, but was apparently not affected by meteorological changes. The examination showed quick reaction of the eye muscles, with spasm of the external rectus; cramp of the masseter muscles on bringing the jaws together forcibly; no involvement of the pterygoids. All the muscles of the upper extremity and of the thorax were involved—indeed, nearly all the muscles of the body. The contraction of the muscles was decidedly tetanic, and was very marked at first, but, on repeated tests, it gradually subsided. The electrical examination showed marked myotonic reaction, and also a wave-like appearance, but it was not certain that this latter phenomenon consisted of a series of waves, such as are observed in water. A piece of muscle had been excised from the biceps, and also from the quadriceps, but they have not yet been minutely examined. The case was quite characteristic on account of the marked variations occurring from time to time. The speaker said that in an article, published by him ten or more years ago, he had taken the stand that these cases were probably of myopathic origin, due to some congenital defect in development, but in the light of modern investigation he was now disposed to believe that some central cause was at work—that there was a functional hereditary derangement of the central nervous system—a condition of lessened resistance in the cells. This did not seem to him a strange assumption, when one considered the well-known idiosyncrasies exhibited to various toxic influences. On the theory that some kind of toxæmia was at the foundation of this disease, he thought the observed phenomena could be explained—at least in this direction seemed to lie the possibility of solving the pathogeny of this class of cases. This patient had not been affected by the disease until eighteen years of age; hence, there was no propriety in calling such a case "*myotonia congenita.*" He would divide these cases into three clsases, viz.: (1) Myotonia congenita; (2) myotonia acquisita; and (3) myotonia transitoria.

DISCUSSION.

Dr. Frederick Peterson asked why a theory of causation might not be founded upon chemical changes in the muscles? Changes in the structure of the muscles, he said, were known to arise—for instance, in connection with typhoid fever.

Dr. Herter thought that we must look to toxic agents as furnishing at least a clew to the causation of such conditions. The peculiar susceptibility to certain types of poisons, seen, for, instance, in epilepsy, must be referred to peculiarities of the central nervous system. He would agree with Dr. Peterson that these cases did not seem to be of central origin, and that it was more probable that they arose from chemical changes in the muscles. To study this subject successfully, it would be necessary to inquire into the condition of the secretions and excretions at the time of the onset of the disease, and not after it had become chronic.

PACHIMENINGITIS HEMORRHAGICA INTERNA IN CHILDREN.

Dr. C. A. Herter said that internal hemorrhagic pachymeningitis was usually considered to be a very rare condition in children, yet one German observer had found it in about 17 per cent. of his autopsies. The following cases were reported:

Case I. A female child, 5½ months old, was admitted to the Babies' Hospital on May 15th, 1897, with an entirely negative family history. The child's illness had begun one month previously with persistent vomiting. The head was of normal shape, and the fontanelles were not bulging. There was a soft spot over one parietal bone. The child had no teeth. On the fifth day after admission tremor and nystagmus developed. Nine days after admission there was a general convulsion, in which the mouth deviated to the left. Cyanosis was a feature of the convulsion. A second one occurred in ten hours. After these seizures the fontanelles were sunken. The child now became semi-comatose, and died after a few days. The autopsy showed the presence of hemorrhagic pachymeningitis, fibrino-purulent pleurisy, pulmonary congestion, fatty liver and nephritis. Along the superior longitudinal fissure, over the entire base and over the island of Reil on both sides was a membrane covering the pia. The ventricles were normal in size, and contained about one drachm of hemorrhagic fluid. There was fluid blood in all the sinuses. The cervical cord showed the same conditions. Under the microscope the right occipital region showed the pia attached to the cortex in many places, and there was a splitting up of the membrane overlying the cortex into two or more layers. The inner layer was infiltrated with small round cells. The outer membranous layers consisted of small round cells, fibroblasts and connective tissue fibres. The island of Reil showed the same condition, but much more marked, and about the same condition was present over the cerebellum. In the spinal cord there were only slight traces of hemorrhage.

Case II. Female infant, colored, 22 months old. The child had been nursed for seven months. It had never walked or stood alone, and was markedly rhachitic. The first two months in the hospital were marked by slight loss in weight and considerable prostration. In October, 1897, the child was re-admitted, with the statement that she had been well up to three days before, at which time she had had four convulsions, followed by three more the next day. The general condition was very bad. The hands and feet were in a position of persistent flexor contraction, characteristic of tetany. The knee-jerks were unobtainable; the fontanelles were bulging. There was slight but varying rigidity of the muscles of the back of the neck. Bloody mucous diarrhœa was present, and the child died in coma. The autopsy showed pachymeningitis hemorrhagica interna, broncho-pneumonia, and acute and chronic ulcerative colitis. Over the right side of the brain was a recent blood-clot covering the entire hemisphere, and over the left occipital lobe. The inner surface of the dura was covered with a membrane extending from the superior longitudinal fissure on either side. The pia was congested. The ventricles and brain substance were apparently normal. All the sinuses were filled with recent clots. The microscopical examination showed thickening of the pia over the right tempero-sphenoidal lobe, and the vessels of the pia were thickened. There was also a thick membrane splitting up into layers, as in the other case. There were numerous small blood vessels, and hemorrhage had occurred into the meshes of the membrane. In places, there were aggregations of small, round cells undergoing fragmentation. They were found chiefly in the superficial layers of the membrane. In the dura the fibres were separated from each other by serous infiltration, and the dura was covered with a membrane similar to that already described. In places, there was very extensive pachymeningitis.

It was at about five months of age, the speaker said, that this disease was especially frequent. The majority of these infants were badly nourished, many of them being subjects of rhachitis or of chronic colitis. The new membrane must be regarded as originating from proliferation of the dural endothelia cells. In some cases there was little inclination to hemorrhage. The membrane was very variable in thickness; sometimes it reached a thickness of two or three lines. It was especially prone to occur in the basal fossæ. There seemed no good reason for thinking that the locality of the pigmentation indicated that the layer of blood originated from the inner surface of the dura. On the other hand, there was no conclusive proof of the old notion that the disease was of inflammatory origin. It was so common to find severe intoxications without such lesions, that the intoxication theory did not seem to him tenable. It was apparently impossible to recognize the condition until the hemorrhage occurred, and even then it was extremely difficult to make a positive diagnosis. Slight cerebral symptoms were probably masked in these very young and usually marantic children. The hemorrhage was probably more often unilateral, and the usual symptoms present were rigidity, hemorrhage and coma. Paralysis was rarely noted. The pyrexia was usually less than in meningitis, but these cases were so commonly complicated with other diseases that the range of temperature was very variable. He did not think there was any symptom or combination of symptoms in hemorrhagic internal pachymeningitis which might not be encountered in any acute infection without any cerebral affection being present; but whenever unilateral rigidity and convulsions, with deepening stupor, were present in a cachectic or rhachitic child under one year of age, we should think of that diagnosis. It was probable that relatively slight traumatisms to the head might occasion rupture of vessels in the highly vascular membrane. This gave these cases a certain medico-legal importance.

DISCUSSION.

Dr. Peterson remarked that the condition was interesting to him because of the possibility of its being found occasionally in infantile cerebral palsy.

Dr. Herter said that he was inclined to think that these membranes were considerably more frequent than one would suppose from the literature. It was quite possible to overlook the presence of the membrane if it were not decidedly vascular.

THE PATHOLOGY AND MORBID ANATOMY OF HUNTINGTON'S CHOREA, WITH REMARKS ON THE DEVELOPMENT AND TREATMENT OF THE DISEASE.

Dr. Joseph Collins said that the neurologist frequently encountered knotty problems, and among these none had the secret of its genesis more carefully concealed than the hereditary degenerative diseases. The pathogenesis of the acute inflammatory diseases of the nervous system was an open book, but the degenerative diseases were discouragingly slow in yielding the mystery of their being. This was especially true of such degenerative diseases as the hereditary ataxias, choreas and dystrophies. The status of the original lesion could not always be inferred from a consideration of the lesion found at the time of death, and this was particularly true if the disease had existed a great number of years. No one could do much laboratory work on the central nervous system of individuals who had succumbed to degenerative nervous diseases of long duration without having forced upon him the fact that there are certain abnormalities of the circulatory system—varying degrees of degeneration of vessels, change in the size of the lymph spaces, and relative disproportion of glia tissue to the parenchyma—which occur with all degenerative diseases, considered entirely apart from their causation. He felt convinced that such changes were very often secondary, and had no other significance than as evidences of protracted disturbances of nutrition, and that this nutritional depravity was the result of the existence of the original lesion. There was nothing more certain than the occurrence of glia proliferation in all slowly progressing destructive lesions of the nervous system, but nothing could be more misleading than to consider this glia overgrowth to be primary, and the changes in the parenchyma secondary.

Huntington's chorea, Dr. Collins said, was a comparatively rare disease, and of rather recent recognition; hence the reports made upon its pathology had not been uniform. The discrepancies were apparently the resultant of the varying points of view of different observers. In studying the nervous system in cases of Huntington's chorea, it was scarcely justifiable to maintain that all the morbid conditions were inherent to the disease, for, as had been said, many of them might be the consequence of prolonged interference with nutrition. A study of two cases, he felt confident, would go far toward establishing the morbid anatomy hinting at the pathogenesis. His patient was a man, fifty-five years of age, who had married in early manhood, and who was the father of three children—all of them giving evidence of neuropathic inheritance. The known duration of the disease in his case was ten years. At the beginnig the hands only were affected, but in the last years the lower extremities were also involved. The mind remained in fairly good condition up to about three years before his death, when he began to have suspicions about his relatives and friends, and became forgetful and suicidal. His speech was so imperfect that in the last years of his life he was understood with difficulty. Dr. Collins had seen him for the first time a few days before his death. He then had a temperature of 105° F., and it remained at about this point until the end. The movements were very severe and incessant, except during sound sleep, although even then they frequently awakened him. He was quite conscious, but made no response to questions. The cause of death seemed to be exhaustion and high temperature. The disease was traceable to the maternal grandfather—an Irishman—who had three children, two of whom were affected with the disease. One of these was the mother of this patient, and of her seven children, five were afflicted with the disease. The other daughter had two children, one of whom became choreic. In three generations there had been no less than nine affected, and when it was considered that many of these children died in infancy, the number of cases that had developed was surprisingly great.

At the autopsy, on opening the skull, the dura was considerably adherent, the diplœ dense and the Pacchionian depressions marked. The brain had a wet appearance, as did also the cord. The pia was not adherent to the brain. The convolution of the anterior portion of the brain were very small, and the entire encephalon weighed 43½ ounces. The dura was intimately adherent to the spinal column. The principal fissures were somewhat wider and shallower and shorter than in the normal brain, but there was nothing pointing to defective convolutions. The average thickness of the gray matter was uniformly less than in the normal brain cortex, but this thinnness could not be attributed here to age. An examination of the pons and medulla oblongata did not show any marked variation from the normal, but the changes were more noticeable lower down. Microscopical changes were not confined exclusively to the Rolandic region, but the process here was more advanced. The specimens were stained by various methods, and carefully examined. The macroscopical changes were briefly as follows: (1) Thinness and atrophy of the cortex; (2) the mottled, streaked appearance and cribriform state on cross section of the brain in the fresh state, due to diminution in number and in health of the ganglion cells and to the increased perivascular and pericellular spaces and increased patency of blood vessels. The microscopical changes were: (1) A decay or slowly progressive degeneration of the ganglion cells of the cortex throughout the brain, especially of the two deepest layers, the layers of large pyramids and polymorphous cells. This cell death was particularly evident in the Rolandic region, very much less so in the anterior pole of the brain, and incomparably less in the posterior pole. (2) Increase of glia tissue, but not sufficiently prominent to constitute sclerosis, the conspicuous increase being about blood vessels and ganglion cells. (3) Enlargement of the pericellular spaces and distention of the pericellular spaces. (4) Slightly diseased blood vessels consisting principally of a proliferation of the nuclei of the adventitia and a thickening of the intima. This involvement of the vessels was not regular or symmetrical, but showed itself in certain sections of vessels only. (5) Relative paucity of the medullated fibres of the cortex. In short, it might be said, that the lesion was a chronic parenchymatous degeneration of the cortex, with consecutive and secondary changes in the interstices, the brunt of the disease having been borne by the motor regions. There was, in consequence, a degeneration of the pyramidal tracts in the spinal cord. In Dr. Dana's case the central convolutions suffered most, and the process occurred in patches throughout the affected cortex. There was nothing to justify the opinion that it was an inflammation—the process was evidently one of degeneration.

In connection with the treatment, Dr. Collins said, that he desired to emphasize the necessity for delaying the advent of the disease in those who had a hereditary tendency to it, and also to emphasize the folly of tenotomy of the eye muscles—a method of treatment now being carried out upon one of these unfortunate individuals in this city, with a promise of a cure. If we wished to inflence the cause of hereditary chorea after it had once become manifest, it would be necessary to administer whatever drug was selected in the largest possible doses consistent with life, and to maintain this medication for a long time.

DISCUSSION.

Dr. Onuf said that he had been present at the autopsy on the case reported in the paper, and had been especially impressed with the general narrowing of the gyri—a general atrophy. On section, the mottled appearance of the cortex had been most striking, bu the cribriform appearance produced by the enlargement of the perivascular spaces was also worthy of note. The general appearance of the brain resembled very closely that of a brain from a case of general paresis. The microscope confirmed the macroscopical appearances, although the changes were not as marked as one would have expected from the gross appearance. The cell changes were of the atrophic order. The characteristic feature was the accumulation of neuroglia cells in the pericellular and perivascular spaces. His impression was, that this accumulation was due to a secondary process following atrophy of the cells. The disease was evidently a degenerative one, originating in the parenchyma of the brain, and not in the interstitial tissue.

Hemiplegia in Whooping Cough.

McKerron (Brit. Med. Jour., Sept. 12th, 1896) reports the following: A child of five and three-quarter years, who had had whooping cough for three weeks, came into the house complaining of a headache and of feeling sick. She vomited once. Right hemiplegia and coma gradually came on, the latter continuing to grow deeper for three days, when it was complete. There were repeated right-sided convulsions. The condition remained stationary for three or four days, and then gradual improvement began, which progressed to almost complete recovery. At the end of three months the right leg was as good as the left, but the child preferred to use the left hand rather than the right.

Patrick (Chicago).

Un Nouveau Signe d'Epilepsie (A New Sign of Epilepsy). Dr. Mairet. (La France Méd., Jan. 29th,'97.)

Numerous experiments prove that the urine of epileptics is hypertoxic before, and hypotoxic after an attack. The hypotoxicity falls from 150 to 450 ccm. (the quantity of urine required to kill a rabbit of one kilogramme), and remains constant during the intervals between seizures. Hypotoxicity is also present in hysterical subjects, but, the urine of epileptics gives rise to convulsive phenomena, whereas that of the former class is simply toxic. In all forms of epilepsy, typical, as well as epilepsy larvata, hypotoxicity exists, which is therefore so characteristic as to assume great clinical and medico-legal importance.

Macalester.

showed that there is a connection of this with the central lobe. Edinger believes the connection is indirectly with the parietal lobe. According to Hösel, Meynert also thought that there is a connection of the nucleus ruber with the parietal lobe. V. Monakow also reports atrophy of the contralateral anterior cerebellar peduncle.

In sections taken from the extreme upper part of the ascending frontal convolution which externally appeared to be normal, as may be seen in the picture, search was made for the giant pyramidal cells (Betz). These cannot be found. Mahaim states that Moeli and Henschen as well as he himself have noted the absence of these cells in cerebral lesions in man.

LA MORT DANS L'HYSTERIE (Death in Hysteria). (Journ. de Méd. et Chir. prat., Aug. 25th, '96.)

Although the manifestations of hysteria are usually considered to be of a benignant character, cases have been observed in which death, due to various conditions, has occurred suddenly. Dr. Le Fournier, in a thesis, has collected a number of illustrative cases, and points to the dangers connected with spasm of the glottis, and anorexia. Thus several cases are reported in which tracheotomy was resorted to, in order to prevent asphyxia. A child, six years old, in imminent danger of being asphyxiated, the attack lasting one hour, was saved by applying the faradic current. Another case was that of a girl, 20 years old, subject to laryngeal spasms, who was revived by artificial respiration, but who died shortly afterwards in a similar attack. Hysterical anorexia ends fatally quite often. After a certain duration the patients fall into a semi-comatose condition, become sleepless, greatly emaciated, and bedridden. If fed artificially in this state the patients continue to lose flesh, the stomach being unable to perform its functions. Death may also occur from incessant vomiting, as for instance: A patient, who was subject to attacks of hysterical vomiting, after taking her evening meal as usual, went to bed and died during the night. The autopsy, made thirty-six hours after death, revealed nothing abnormal of note, and the stomach contained four ounces of food. Occurrence of sudden death in hysterical subjects has further been observed to be due to attacks of angina pectoris, and supervening upon abdominal operations, especial vaginal hysterectomy. MACALESTER.

lesions of this nerve and concluded, therefore, that this nucleus must be motor. Van Gehuchten states that he has found changes in the acoustic nucleus of the oblongata a short time after section of the eighth nerve. The axis cylinders which pass into this nucleus, *terminate* about its cells. He believes that changes occur in the cell body of the central neuron after lesion of the peripheral neuron. If we dare believe that neurofibrils pass from the first neuron into the cell body of the second, we can readily understand why there should be an alteration of structure in the latter.

Let us look at the subject of the reflexes in the light of Apáthy's discoveries. If it is true that neurofibrils pass from sensory fibres into motor cells, we may be able to better understand the difficult subject of the reflexes. It is not unusual in great exaggeration of the reflexes to obtain contraction of the adductor muscles of one thigh by striking the patellar tendon on the opposite limb. A reflex is not limited to one or even a few muscles. If neurofibrils pass through a number of ganglion cells, we may readily understand how an impulse acts on many cells.

Dare we believe that the many neurofibrils which leave a sensory cell, in contrast to the single one which leaves a motor cell, are indicative of the conveyance of different forms of sensation?

These are only a few thoughts which occur to me. I am seeking light, not stating facts, and am well aware of the danger of theorizing, but time will demonstrate the truth or fallacy of these suggestions.

NEW AESTHESIOMETER.

In the June number of the Bulletin of the Johns Hopkins Hospital, Dr. L. F. Barker describes an ingenious little instrument recently prepared by Professor Von Frey for studying pain and pressure sense. It is an improvement on an apparatus previously described from Von Frey's clinic by Dr. Barker, in which the amount of pressure necessary to bend test hairs of various diameters was employed. The new instrument has the advantage that with a single hair one can obtain a number of different pressure values.

It consists of a long hair pushed through a capillary tube like a thermometer tube; by sliding a sheathing over the tube it may be made to project more or less. According to the length of the projection of the hair is the strength which is necessary to make it bend, and a reading of the scales inscribed upon the tube gives thus some idea of the force used. Dr. Barker quotes a case in evidence of its usefulness, in which ordinary slight stimuli appeared to cause pain. It was supposed that pressure sense was absent. It was easy with this instrument to prove that pressure sense was not abolished, though the level for pain was almost the same as that for touch. The importance for such an observation is obvious for showing the retention of tactile sense in cases where it is obscured by hyperesthesia. MITCHELL.

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